

Spontaneous Coronary Artery Dissection in a Patient with Systemic Lupus Erythematosus

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Abstract

Spontaneous coronary artery dissection (SCAD) is an uncommon condition that may lead to sudden coronary artery occlusion resulting in a fatal acute myocardial infarction. It usually affects young to middle age women. A Medline search from 1966 to 2001 (using keywords: coronary artery dissection and systemic lupus erythematosus) revealed no prior reports of coronary dissection in a patient with systemic lupus erythematosus (SLE). We describe a 48-year-old woman with SLE who sustained a fatal spontaneous left main coronary artery dissection. Coronary angiogram was notable for marked variability in the size of coronary lumen from systole to diastole. This case demonstrates the need to consider SCAD in the evaluation of chest pain and myocardial infarction in patients with SLE. Furthermore, in the absence of classical angiographic findings of coronary dissection, a detailed review of phasic changes in coronary lumen during a cardiac cycle could help reach this diagnosis.

Introduction

Spontaneous coronary artery dissection (SCAD), an infrequent cause of acute myocardial infarction, was first reported by Pretty¹ in 1931. More than 200 cases have been reported in the English literature. More than 80% of these occur in women 20–50 years old. SCAD has been reported to occur in association with pregnancy, oral contraceptives,² intense physical exertion,³ blunt chest trauma,⁴ mitral stenosis,⁵ and connective tissue disorders like Ehlers-Danlos syndrome.⁶

A Medline search from 1966 to 2001 (using keywords: coronary artery dissection and systemic lupus erythematosus) revealed no reports of coronary dissection in a patient with systemic lupus erythematosus (SLE). In this report we describe a 48-year-old woman with SLE who sustained a fatal spontaneous left main coronary artery dissection.

Case report

A 48-year-old white female with a 15-year history of SLE and hypertension was referred to our institution for further management of acute infero-posterior myocardial infarction. On the day of admission she awoke with severe, crushing retrosternal chest pain associ-

ated with diaphoresis and shortness of breath. Her SLE, currently quiescent, had been manifested primarily by arthritis and skin lesions. At the time of presentation she was taking hydroxychloroquine sulfate. There was no history of diabetes mellitus, hyperlipidemia, smoking, use of oral contraceptives, recreational drug abuse or family history of accelerated atherosclerosis.

Initial physical examination revealed an alert, anxious middle-aged female with blood pressure of 134/76 mmHg and regular pulse rate of 125/min. Cardiovascular exam was normal. There was no carotid bruit. Peripheral pulses were normal and symmetrical. Chest roentgenogram was normal. The electrocardiogram showed an acute infero-posterior myocardial infarction. In the emergency department, she was treated with aspirin, intravenous nitroglycerin, heparin, metoprolol and tenecteplase without resolution of chest pain or ST-segments. Upon transfer to our facility, she developed cardiac arrest with pulseless electrical activity. Resuscitation was performed successfully.

Emergent cardiac catheterization revealed a normal left main coronary artery. The mid-segments of left anterior descending artery (LAD) and left circumflex artery were occluded. The right coronary artery was normal. The guidewire could not be advanced across the occlusion in left circumflex. The LAD occlusion was treated with a 3 x 18 mm stent. During the procedure she developed complete heart block and hypotension that necessitated insertion of a pacing Swan-Ganz catheter and an intra-aortic balloon pump. Two hours later, the patient had a second cardiac arrest and expired.

At autopsy the heart weighed 420 grams and showed mild dilatation of both atria, and left ventricle. Multifocal extensive contraction band necrosis involving the interventricular septum, anterior wall, and posterior wall was present. The coronary arteries showed an extensive dissection that compromised the lumen of left main, left anterior descending, and left circumflex arteries. The dissection plane was between the outer third of media and extended to the adventitia (Fig 1).

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Figure 1.— Autopsy slides of coronary dissection

Panel A shows extensive contraction band necrosis (arrows) involving the antero-lateral wall. Dissection involving the left main coronary artery (Panel B), and the left circumflex (Panel C) are shown. Panel D shows the stented segment of the left anterior descending coronary artery with a patent lumen.

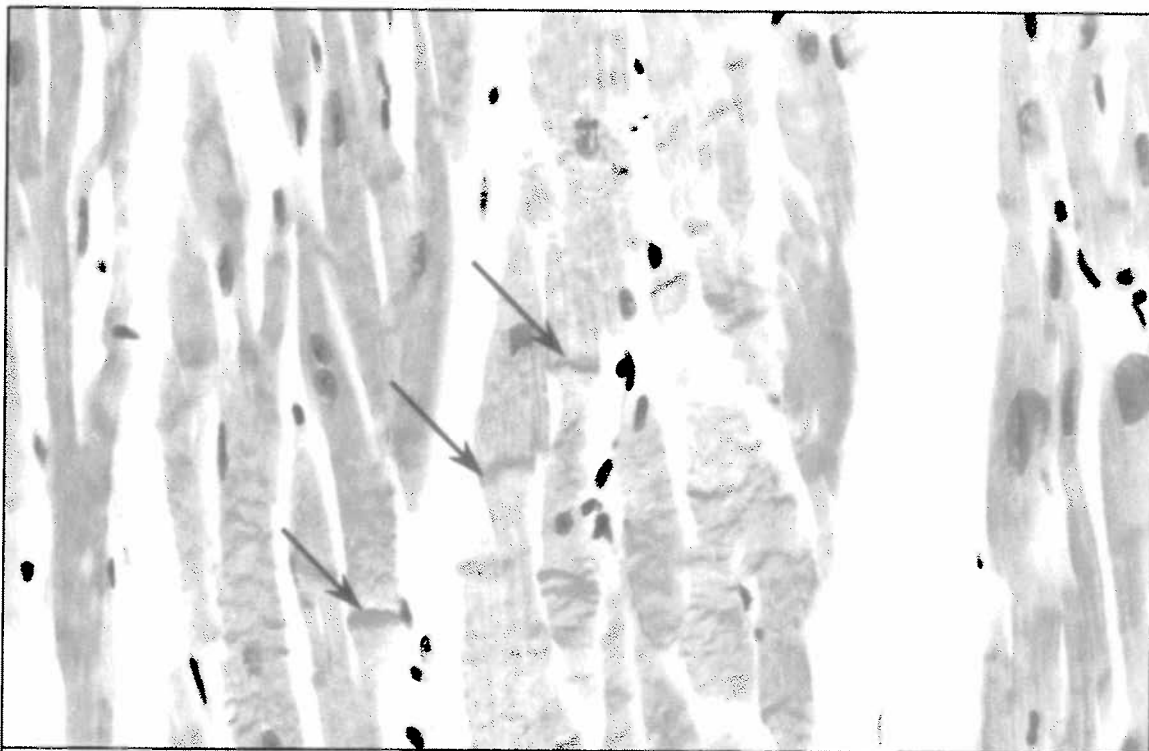


Figure 1, Panel A

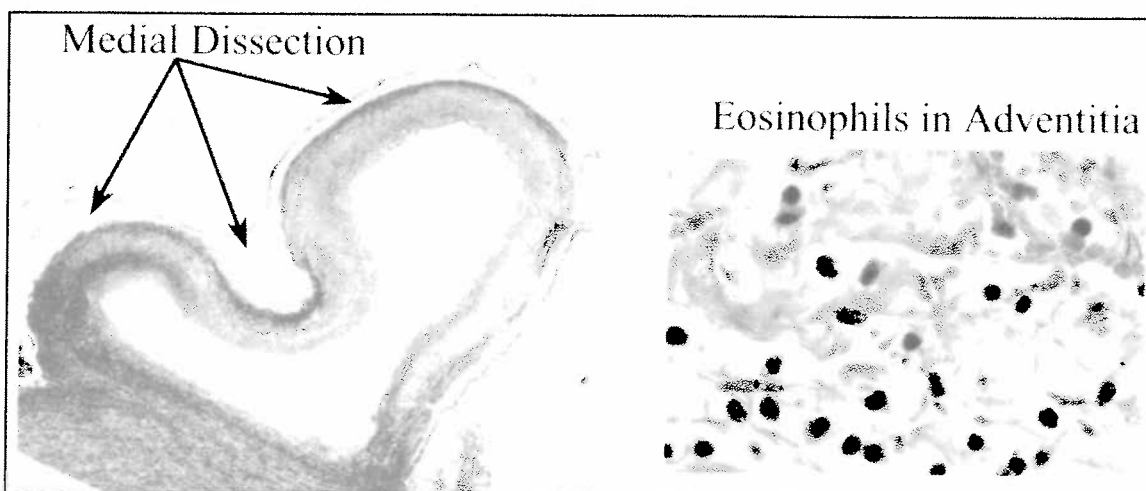
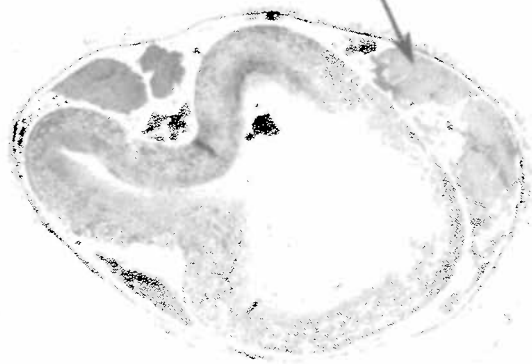


Figure 1, Panel B

Circumferential Arterial
Dissection



Platelet-Rich Thrombus +
Acute Inflammatory Cells
In Dissection Plane

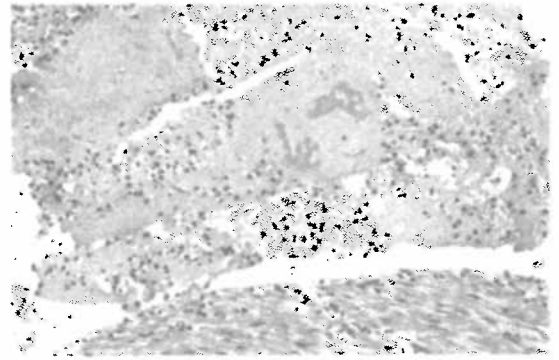
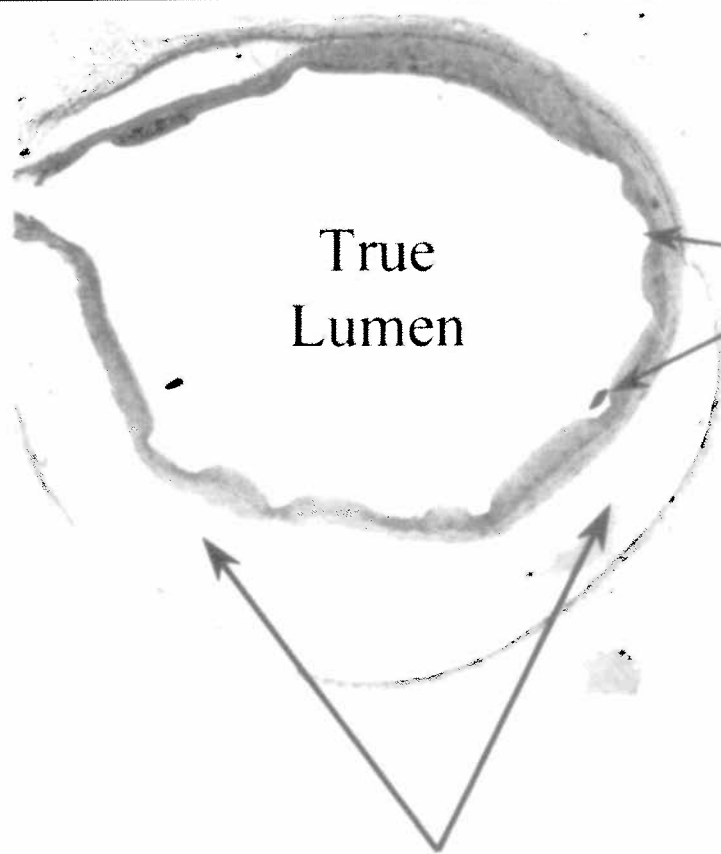


Figure 1, Panel C

True
Lumen



Stent Struts

Compressed Dissection Plane

Figure1, Panel D

The lumen at the site of stent was widely patent and the medial dissection was effectively compressed. Platelet-rich thrombus with acute inflammatory cells was present in the coronary dissection plane. Numerous eosinophils were noted in the adventitia of the left main and left main bifurcation.

Postmortem review of the coronary angiogram showed an unremarkable left main and right coronary

arteries, with occluded mid-segments of LAD and left circumflex arteries. There was no intimal flap or extraluminal contrast dye. Careful evaluation revealed that the proximal segment of LAD expanded fully in diastole (Fig 2A) and collapses in systole (Fig 2B). Similar changes in the coronary lumen were also seen in the left circumflex artery.

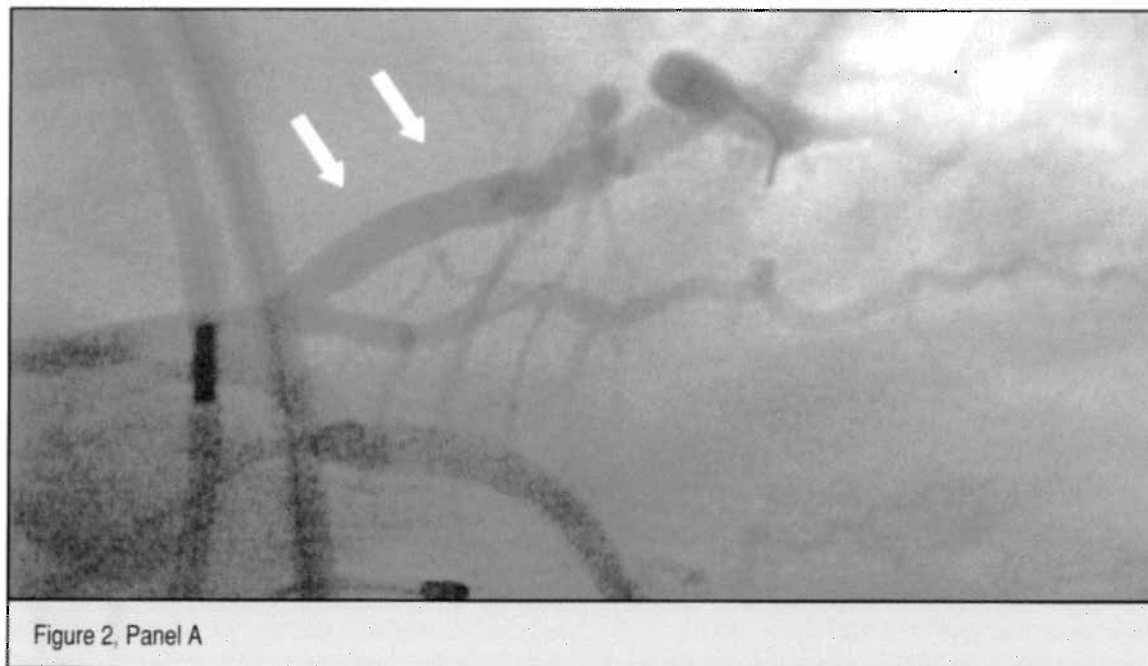
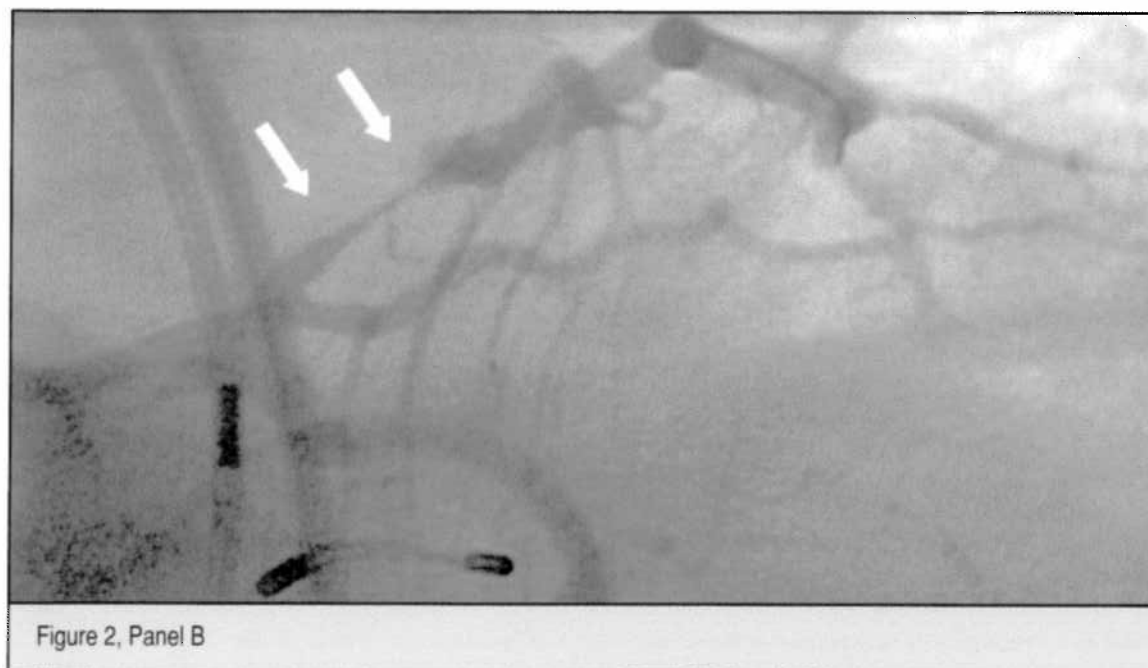


Figure 2.— Angiogram of the left coronary artery while on intra-aortic balloon pump

Panel A shows a diastolic frame with adequate expansion (white arrows) of the coronary lumen of proximal LAD. Panel B shows a diffuse collapse of the proximal LAD lumen during systole while adequate contrast dye and expansion is noted in the segments proximal and distal to this lesion. There is no extravasation of contrast material.



Discussion

This is the first reported case of spontaneous coronary artery dissection in a patient with SLE. SLE can affect all parts of the heart, including the pericardium, myocardium, valves, conduction system and the coronary arteries.⁷ More than 50% of SLE patients may have cardiac involvement with 25-45% having coronary artery disease.⁸ The prevalence of coronary artery disease in SLE patients is at least 50-fold greater than among age-matched controls.⁹ Clinical manifestations of coronary artery disease in SLE can result from several pathophysiological mechanisms, including accelerated atherosclerosis, arteritis, thrombosis, embolization, spasm, and abnormal coronary reserve.⁸ While it is possible that SLE was a coincidental condition in this case of SCAD, the latter should be added to the list of SLE-associated coronary complications.

The true incidence of SCAD is probably underestimated, as most of the cases are diagnosed at autopsy.¹⁰ The LAD artery is involved in 80% of cases, while right coronary and left circumflex arteries are involved less frequently. Approximately 15% of patients present with involvement of left main coronary artery.¹¹ Multiple vessels could be involved in SCAD at initial presentation.¹²⁻¹⁴ In our case, the dissection extended from left main coronary artery to both LAD and left circumflex arteries. This type of dissection is rare,¹¹ and associated with high mortality. In men right coronary dissections are more common (73%), while in women left coronary dissections are reported to occur more frequently (88%).¹⁰

Angiographic findings in dissection include coronary occlusion, thrombus formation, presence of intimal flap and persistence of contrast dye in the false lumen. When neither an intimal flap nor a false lumen is recognized, a dissection could be suspected by a careful review of the angiogram during the different phases of the cardiac cycle. In diastole, a lower distending pressure in the true lumen compared to the false lumen leads to the collapse of true lumen along the dissected segment. In systole, when the coronary artery pressure in the true lumen exceeds the pressure in the false lumen, the relative luminal diameter is larger. In patients with an intra-aortic balloon pump, the coronary perfusion pressure is highest in diastole. Therefore in patients with intra-aortic balloon such as ours, this phenomenon of phasic changes of coronary lumen in systole and diastole is reversed. Of note, the NHLBI classification¹⁶ of coronary dissection with subtypes A to F does not describe these phasic changes in relation to the true and false lumen size during the cardiac cycle. However, this classification was intended for post-procedural dissection and not for SCAD. These dynamic changes are likely seen early in the course of dissection when either the dissection plane has not completely thrombosed or the dissection

is still expanding. Phasic changes in coronary lumen are also seen in "myocardial bridging," however, these changes are often not seen in proximal LAD. Furthermore, unlike SCAD the collapse of the lumen occurs in systole rather than diastole.

The exact etiology of SCAD is still uncertain. The increased incidence during the peripartum period and the observed histological changes in blood vessel wall¹⁷ suggests the potential role of hormonal changes and hemodynamic stresses. A periadventitial infiltrate composed of eosinophils has been noted in patients with SCAD. It is postulated that spontaneous dissection results from an accumulation of eosinophils that secrete lytic enzymes and major basic protein, leading to medial weakening.¹⁸ Others, however, believe that inflammation is a consequence rather than the cause of dissection. Another proposed mechanism is disruption of vasa vasorum leading to intramedial hemorrhage and subsequent dissection without an intimal tear.¹⁹ In patients with SLE, fibrinoid necrosis and inflammation of the media could be contributory; however no evidence of vasculitis was seen in our patient.

The prognosis of patients with spontaneous coronary dissection is poor. About 50% of early reported cases have been diagnosed postmortem. In 1991, Benham and Tillinghast²⁰ reviewed 123 cases of spontaneous dissections and reported a 67% mortality rate. Tsimikas et al.²¹ in a review of SCAD that occurred after 1993, reported an improved survival (78%). Improved prognosis is observed with early diagnosis, aggressive medical therapy or revascularization therapy, and in the absence of left main involvement.

Various treatment options have been suggested for SCAD; however, there is no agreement on the optimal therapy. Spontaneous healing of a coronary dissection has been reported.⁵ Thrombolytic therapy has been successfully used in SCAD; however, due to the potential risk for propagation of dissection and expansion of the intramural hematoma the use of thrombolytics should be avoided.²²

Nonsurgical revascularization with primary angioplasty or intracoronary stenting offers a more definitive treatment for single vessel SCAD. Successful use of angioplasty has been reported.²³ Fish et al.²⁴ described the first use of intracoronary stenting, and since then additional cases of successful coronary stent implantation have been reported.²³ Stent implantation offers superior efficacy to angioplasty alone, as stents are better able to mechanically compress the medial dissection plane. Coronary artery bypass graft (CABG) has been successfully performed in patients with SCAD,²⁵ and heart transplantation has been done in cases of severe heart failure.²⁶

Overall, the optimal treatment of spontaneous coronary dissection depends on the patient's clinical status and location and extent of the dissection. Medical

therapy is appropriate for asymptomatic patients with non-occlusive disease and good distal flow. For symptomatic patients with single vessel involvement, primary stenting is indicated. CABG should be strongly considered in patients with left main coronary artery or multivessel dissection.

This case demonstrates the need to consider SCAD in the evaluation of chest pain and myocardial infarction in patients with SLE even in the absence of classical angiographic findings. Attention to phasic changes in the coronary lumen may be helpful in the diagnosis of SCAD.

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